Endocarditis of an ASD Occluder Mimicking Adult-onset Still Disease in a Grown-up with Congenital Heart Disease

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A 40-year old male patient was admitted with the diagnosis of endocarditis of an atrial septal occluder via the spine surgery of a local hospital. The occluder was implanted 2001 for a detected atrial septal defect type 2. In the admitting hospital the patient presented himself with spondylodiscitis and ongoing recurrent fever episodes and was scheduled for surgical treatment of the focus. Transthoracic and transesophageal echocardiography done before surgery revealed the diagnosis of endocarditis. In conjunction with the result from blood cultures showing staphylococcus aureus, antibiosis with flucloxacillin, gentamicin and clindamycin was started according to the antibiogram.¹

In the previous medical history the patient had a tooth extraction 3 month ago under concomitant medication with corticosteroids, methotrexate and anakinra due to a diagnosed adult-onset Still disease (AOSD), antibiosis was not given as prophylaxis as the duration since implantation was longer than 6 month. Typical symptoms of morbus Still were present with a salmon-colored rash, fever and pain in several joints being diminished by the medication in the past history. On initial assessment with transesophageal echocardiography two vegetations were visualized with a diameter of 11x10cm and a second vegetation of 6x7cm (figure 1A and video 1) on the right atrial side of the occluder. After discussion of the case in the heart team with the cardiac surgeons and the immediate clinical improvement and vanishing fever as response to antibiotic treatment, urgent surgical treatment was postponed. After a period of five days of treatment the control transesophageal echocardiography (figure 1B+C and video 2) could not show the vegetation on the occluder and there were no signs of distal embolisation into the lung circulation. The patient was treated for four weeks with the antibiotic regime and was without symptoms after ending the treatment. After one week without

Figure 1. A-C. (A) A second vegetation of 6x7cm (pictured with red arrows) on the right atrial side of the occluder, (B) and (C) transesophageal echocardiography no longer showing the vegetation on the occluder.

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treatment blood cultures were without proof of staphylococcus aureus and the transesophageal echo was without a sign of recurrence of the endocarditis.

After a period of 6 month the patient presented without any sign of the diagnosed AOSD and immunosuppressive therapy had been ended. The diagnosis of AOSD was 6 months before the tooth extraction and it was suggested that symptoms could have been due to an occult infection before, aggravated by immunosupression resulting in endocarditis of the occluder and the spondylodiscitis. However, ASD occluder endocarditis is very rare and uncommon after the first 6 month after implantation and without residual shunt, most often surgery is recommended. Although the combination of symptoms in the first place suggested AOSD, prosthetic material, like the ASD occluder, should trigger the search for signs of endocarditis, especially in grown-ups with congenital heart disease. The concomitant immunosuppressive therapies lead to a smoldering course resulting in multiple external manifestations of endocarditis which were attributed to AOSD in the first place. Grown-ups with congenital heart disease are at a high risk of endocarditis even after treatment and any infection without focus should be investigated by an “endocarditis team” as suggested.

Conflicts of interest
The authors declare no conflicts of interest.

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